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Imaging in ACHD-PAH – What the PH/CHD Specialty Center Offers

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Dr. Krasuski:
Imaging in the ACHD-PAH patient. What the PH/CHD Specialty Center Offers. So, imaging of the congenital heart disease patient is quite challenging. You identify what your goal should be. Ideally, you should determine the original and current or post-op anatomic relationships. First step often in these patients is to get the old medical record to know exactly what was done for the patient. If those records are not available, try to get as much from clinical notes, simple studies. But if you don't have that information, imaging can be very important. You want to assess for residual lesions, things such as shunts. We've talked about ASDs, VSDs, PDAs, collateral vessels. You want to look for valvular heart disease, either stenosis or regurgitation. You want to look for vascular obstructions in pulmonary arteries and veins, right and left ventricular outflow tracts. You want to measure differential blood flow to the two lungs. You want to assess for the ventricular size, thickness and function. Not only for the subpulmonic chamber, but also for the systemic chamber. And you want to appraise myocardial injury. We now know that myocardial scar is a very important predictor of long-term clinical outcome, particularly the risk for arrhythmia and sudden death.

Now there are different modalities available. Echocardiography really, I think, are often first step in the toolbox. It's non-invasive, there's no radiation, it's readily available, we don't need to sedate the patient, we can visualize the intracardiac anatomy beautifully, and it provides good temporal resolution. Unfortunately, not every patient is well-assessed with this modality. Some patients have lung disease or they're obese and they have poor acoustic windows. Also, a lot of scar tissue can impact the ability to visualize the cardiac structures. There's also poor visualization of the extracardiac anatomy. Structures such as the aorta, or the descending portion of the aorta, can be often limited in its assessment, Nuclear perfusion imaging can allow us to quantify pulmonary blood flow, but it really does provide very limited information and does subject the patient to radiation. CT scanning is noninvasive, has excellent spatial resolution, but again, there's radiation, there's a need for contrast dye which can increase the risk for the development of renal dysfunction. And then there's limited intracardiac anatomy, and there's no hemodynamic data oftentimes for these patients, where the hemodynamic data is not as carefully assessed as it is with CMR. So CMR is noninvasive, there's no radiation. It can assess the intra and extracardiac anatomy, ventricular function, myocardial viability, hemodynamic data, lung perfusion. Sounds like the perfect modality. The problem is it's expensive, it's not readily available, it's time-consuming, you get metallic artifact. And some devices, pacemakers, ICDs, are still contraindicated to place into the scanner. Often there's abandoned leads, pacemaker leads and ICD leads. Those are still a contraindication. Angiography can be performed. This provides excellent visualization of the extracardiac anatomy. You can also get hemodynamics and facilitates intervention, but this is the most invasive. There's a need for sedation in many of these patients, radiation, and there's poor visualization of the intracardiac anatomy.

So here is just simple echocardiography. This is a very complex patient with congenital heart disease. You can see some of the limitations here. You get a lot of artifact in some of these images, but you do have the ability to use Doppler and assess pulmonary pressures, you can assess systemic chambers and gradients across valves, so it can be very useful from a hemodynamic standpoint.

But again, catheterization in some of these patients is the most accurate pressure measures.

Here's an example of a nuclear perfusion scan. This is a very helpful study when we were looking for thromboembolic disease, but it can also allow us to quantify the blood flow to the lung segments, and this can be helpful particularly in patients with segmental pulmonary hypertension and peripheral pulmonary stenosis where we look at variable lung perfusion. CMRI provides the most accurate way, or CT is another way to do this. The pictures are beautiful, but again, an MRI requires usually about an hour for a patient. There's a lot of breath-holding. It can be quite the challenge for a patient to do this. And again, availability is an issue. CT is much quicker but requires radiation and contrast time. Here is a pulmonary angiogram for a patient with very distorted peripheral pulmonary circulation here. It can provide very nice data, but it is invasive. There can be complications related to this, but it does allow us to get position and to perform interventions.

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